International Journal Dental and Medical Sciences Research
Volume 3, Issue 5, Sep-Oct 2021 pp 1194-1198 www.ijdmsrjournal.com ISSN: 2582-6018

A rare case of subserosal uterine leiomyosarcoma in a perimenopausal women - A case report

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Submitted: 15-10-2021 Revised: 28-10-2021 Accepted: 30-10-2021

ABSTRACT:Uterine leiomyosarcoma is a malignant tumor of the smooth muscles of the uterus. It is an aggressive malignancy and has a high mortality and morbidity. This tumor can easily be misdiagnosed as leiomyoma of the uterus, a benign condition and most of the times are diagnosed postoperatively through histopathological examination. Here we present a case of a 47 year old female P5L5, perimenopausal who had complaints of abnormal uterine bleeding, abdominal pain and mass per abdomen for 1 year. The patient was diagnosed with Fibroid uterus and proceeded with total abdominal hysterectomy with bilateral salpingo oophorectomy. The specimen was sent for histopathological analysis that revealed uterine leiomyosarcoma. She was referred to the tumor board for deciding on further policy of management and metastatic work up showed no distant metastasis. However due to rare occurrence, advances remain limited and surgery still remains the mainstay of management. Preoperative diagnosis of leiomyosarcoma is difficult and MRI of the pelvis is recommended to diagnose preoperatively.

KEYWORDS:uterine leiomyoma, uterine leiomyosarcoma , uterine cancer, leiomyosarcoma

I. INTRODUCTION

Uterine leiomyosarcoma is the malignancy of the smooth muscles lining the walls of the uterus. It is an uncommon tumor and it accounts for only 8 % of all uterine malignancies(1) .The usual presentation of leiomyosarcoma is abnormal uterine bleeding , pelvic pain and symptomatic mass effect. These features are also common to benign leiomyoma of the uterus, which is a common condition(2-4). Due to an overlap among the symptoms between a leiomyoma and a leiomyosarcoma and the rare occurrence of a leiomyosarcoma, preoperative diagnosis is seldom possible and needs a high index of suspicion. Though a rapidly growing uterus is highly suspicious of sarcoma the review of literature does not support this as the occurrence of sarcoma in patients who are operated for leiomyoma that is rapidly growing is around 0.27%(5). Therefore, it is usually diagnosed only after a surgery on histopathological examination as there is no reliable diagnostic test. Here we present a case report on a 47yearold perimenopausal who was diagnosed as fibroid uterus preoperatively and postoperatively biopsy report showed leiomyosarcoma.

II. CASE REPORT

A 47-year old female, perimenopausal, P5L5 from Katpadi, Tamil Nadu came with the chief complaints of abnormal uterine bleeding, abdominal pain and mass per abdomen. She had been experiencing excessive bleeding per Vagina for the past 1 year and was associated with dysmenorrhea. Abdominal pain was dull aching, constant and was radiating to the back. Mass per abdomen for 1 year, in the lower abdomen, insidious onset, gradually progressive initially and rapid progression over the last 8 months. No history of loss of appetite or loss of weight. No history of similar complaints in the past. Patient had undergone postpartum sterilization 19 years back. No other major abdominal operation. There is no history of prior pelvic radiotherapy.

On examination patient was conscious, oriented, stable vital signs and mild pallor examination of the abdomen showed that the patient had a 15×10 cm occupying the umbilicus and the suprapubic region extending partly into the right and left iliac fossa with a size corresponding to 20 weeks, Not tender, Surface of the mass was found to be bosselated. The lower border of the mass cannot be made out and it had side to side mobility. On bimanual palpation transmitted mobility was present and there was no cervical motion tenderness.

International Journal Dental and Medical Sciences Research
Volume 3, Issue 5, Sep-Oct 2021 pp 1194-1198 www.ijdmsrjournal.com ISSN: 2582-6018

On further investigation with an ultrasonogram of the abdomen it showed a 11.5×11 cm lesion arising from the uterus suggesting a submucosal fibroid. Endometrial biopsy showed normal study and VIA/VILI was found to be negative. Contrast enhanced computed tomography of the abdomen was done and it showed a large and exophytic heterogeneously enhancing mass arising from the serosal portion of the fundus and the proximal body of uterus with internal enhancing solid components. Ovaries not visualized separately and an impression of subserosal fibroid of the uterus was arrived (Figure 1).

A provisional diagnosis of a sub serosal leiomyoma of the uterus was made and we proceeded with total abdominal hysterectomy with bilateral salpingo-oophorectomy. Intraoperatively - A 20×15 cm fibroid arising from the fundus of the uterus predominantly on the right side and another fibroid of 5×4 cm was present inferior to it (Figure 2). The resected specimen was sent for histopathological analysis concluded the final diagnosis of leiomyosarcoma of the uterus.

Grossing of the specimen showed a mass of size 20×14×9 cm subserosal lay with dilated veins on its surface and cut section of the uterus revealed - endometrial thickness of 0.3 cm and cut section of the mass showed -grey white whorled areas with hemorrhagic and cystic degeneration. Microscopic examination of the tumor showed cellular neoplasm dispersed in follicles and interlacing bundles composed of spindle shaped smooth muscle cells with scant eosinophilic cytoplasm nuclei exhibiting diffuse atypia. Pleomorphic bizarre tumor giant cells are also seen mitotic index of 5/10 High Power Field (HPF) was noted and an impression of leiomyosarcoma of the uterus - high grade tumor.

On follow up visit - examination of the abdominal wound and the vaginal vault was found to be healthy and patient was presented in tumor board for deciding on further plan of management and adjuvant pelvic radiotherapy was recommended and patient referred to department of radiation oncology for pelvic radiotherapy.

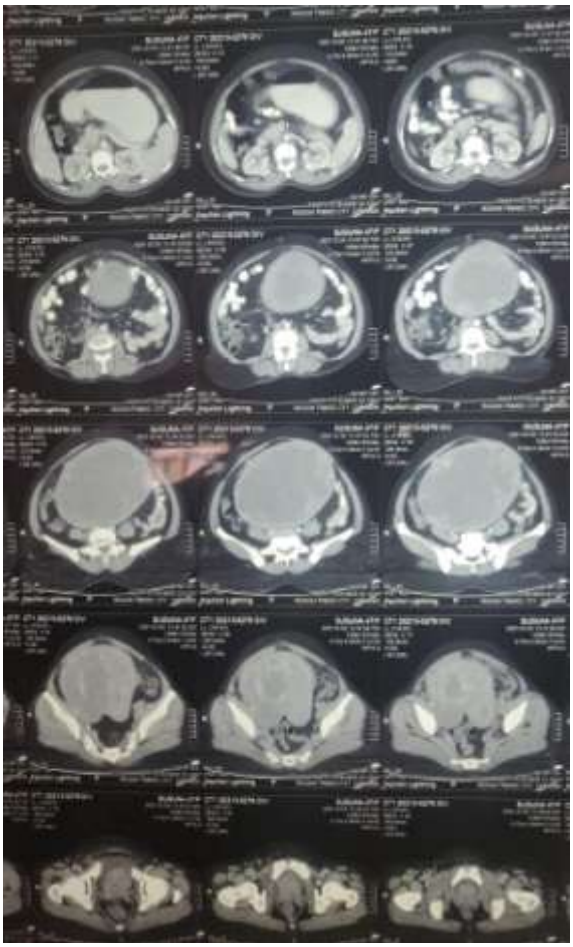

Figure 1. CECT images showing the abdominopelvic mass arising from the uterus

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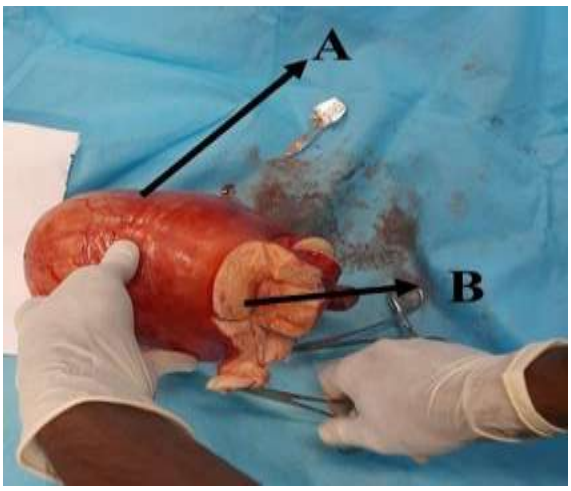

Figure 2. shows the cut section of the resected specimen that includes the uterus, cervix and both the ovaries. (A) shows the subserosal tumor (B) showing the uterine cavity

III. DISCUSSION

Uterine leiomyosarcoma is an uncommon malignant lesion of the uterus and it accounts for about 8 % of malignancies of the uterus(1) and 0.5% of the sarcomatous transformations in a benign leiomyoma(6).It is the most common of the uterine sarcomas. Prior pelvic radiotherapy and Tamoxifen are considered as risks for uterine carcinosarcoma but not for leiomyosarcoma(9,10,11) . The clinical features of a leiomyosarcoma includes abnormal uterine bleeding , mass lesion per abdomen and pelvic pain(2-4). In a postmenopausal women it also includes postmenousal bleeding and an increasing size of a previously diagnosed fibroid. Leiomyosarcomas are usually diagnosed around the 5th and 6 th decade of a woman's life(7) . A Rapidly enlarging leiomyoma though always raises the suspicion of a leiomyosarcoma, the literature doesn't support this theory as the incidence of a leiomyosarcoma for a preoperatively diagnosed as a leiomyoma is only 0.5 %(6) . In addition there is also an overlap among the clinical features of a leiomyoma and leiomyosarcoma , which makes the preoperative diagnosis of the latter more difficult and there is a lack of a reliable diagnostic method for the diagnosis of the same. Hence the diagnosis of a leiomyosarcoma is usually done after a surgical procedure on histopathological examination.

On histopathological examination leiomyosarcoma is usually diagnosed with nuclear atypia, mitotic figures and coagulative necrosis(8,17). Presence of any two of the three is diagnostic of leiomyosarcoma. Preoperative evaluation of uterine neoplasms with biochemical, radiological imaging or endometrial sampling had not given a sufficient result in absolutely excluding the diagnosis of leiomyosarcoma(12-16) . Endometrial biopsy evaluates the endometrial lining of the uterus and can give some information if the lesion is submucosal , and is not useful if the lesion exists in other planes. Serum lactate dehydrogenase and Magnetic Resonance Imaging (MRI) are being evaluated as potential tools of investigation for the diagnosis of leiomyosarcoma but have not been sufficiently tested(12,13,18,19).

The treatment of uterine leiomyosarcoma is total hysterectomy. Surgery is the mainstay of treatment and to ensure complete excision of the primary lesion and to avoid tumor spillage hysterectomy is the standard of care and morcellation of the uterus during hysterectomy to be avoided. Ovarian preservation in a nulliparous women with uterine leiomyosarcoma is controversial because there is a potential for hormonal stimulation via estrogen receptor of residual disease and that the withdrawal of estrogen appears to lead a reduction in tumor growth and a case report also suggests the regression of metastatic uterine smooth muscle tumor after removal of the ovaries . Therefore ovarian preservation may adversely impact survival. However further studies have reported an equal rate of recurrence in both the groups of women who had undergone oophorectomy and those who had ovarian preservation(24,25). Ovarian preservation can be a reasonable option in women who want to preserve fertility with no gross extrauterine disease(20,21,22,23)

Recent advances in the treatment options in uterine leiomyoma also complicated the diagnosis of leiomyosarcoma as there is no tissue diagnosis in Uterine Artery Embolisation (UAE) .

Though the incidence of leiomyosarcoma is small , there is a large risk of delay in the diagnosis of a condition that is associated with high morbidity and mortality. Hence in women undergoing UAE failure of the tumor to respond or rapid return in symptoms should raise the suspicion of a leiomyosarcoma and it should not be recommended for postmenopausal women with fibroid.

Adjuvant pelvic radiotherapy is usually reserved only for palliative care, as there had been no difference among the groups that did and did not receive pelvic radiotherapy though improved locoregional control had been demonstrated in others(25,26,27) . It's been postulated that sarcomas usually metastasise hematogenously and recurrence at distant sites and hence no benefit had been observed with pelvic radiotherapy in clinical outcome. (27)

In discussion regarding chemotherapeutic agents,doxorubicin is effective against uterine leiomyosarcoma and is in use . However the introduction of gemcitabine and docetaxel as first and second line agents respectively had demonstrated significant effect against the tumor. Temozolamide has also been tried against recurrent uterine leiomyosarcoma(28) .

Molecular assays had demonstrated that Matrix MetalloProteinases (MMP) are involved in tumor invasion and metastasis.The expression of MMP -1 (86%) is more than MMP 2 (48%) in uterine leiomyosarcoma. MMP 2 expression is correlated with vascular invasion and tumors negative for MMP 2 are associated with better disease-free survival. (29)

Secondary cytoreduction for isolated pulmonary metastasis with resection is associated with overall survival of 45% and 35% at 5 years and 10 years respectively (30). The 5-year survival rate of leiomyosarcoma ranges from 25-75%. Prognostic factors include tumor grade, stage of the tumor and mitotic count. Absence of vascular space involvement was associated with enhanced survival. (24,25,26,27)

IV. CONCLUSION

High suspicion is required for preoperative diagnosis of leiomyosarcoma. In high risk cases , MRI of the pelvis is recommended. Surgery is the mainstay of treatment and morcellation of the tumor to be avoided to decrease tumor spillage and thereby metastasis of the tumor. Furthermore, molecular studies are essential for development of newer chemotherapeutic agents to increase the survival of the patient. Uterine artery embolization when used in treatment of fibroids , and the tumor fails to respond or there is a rapid recurrence of symptoms of leiomyosarcoma to be suspected.

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